Cognitive Findings in Benign Childhood Epilepsy with Occipital Paroxysms

Oksipital Paroksizmin Eşlik Ettiği Benign Çocukluk Çağı Epilepsilerinde Kognitif Bulgular

Neurocognition in Occipital Epilepsy

Ebru Kolsal[1], Ayse Serdaroglu[2], Sebnem Soysal[2], Esra Gurkas[2], Unsal Yılmaz[2]
[1]Bakirkoy Dr. Sadi Konuk Research Hospital, Department of Pediatric Neurology, İstanbul,
[2]Gazi University Faculty of Medicine, Department of Pediatric Neurology, Ankara, Türkiye

Özet

Giriş: Bu çalışmada iyi huylu çocukluk çağı oksipital epilepsilerinde görsel algılamayı ve kognitif fonksiyonları değerlendirme hedeflenmiştir. Gereç ve Yöntem: Çocuk nöroloji polikliniğine 2009-2010 yılları arasında başvuran 1-18 yaş arası hastaların hastane kayıtları değerlendirildi. Benign oksipital lob epilepsi tanısı olan 21 hasta belirlendi. Bu hastalardan çalışmaya katılmayı kabul eden 16 hasta nöropsikolojik testlerle değerlendirildi. Bulgular: Denver gelişim testi ile değerlendirilen 5 hastadan 2 tanesi yaşlarına göre daha düşük puanlar aldılar. Kalan 11 hasta Wechsler zeka ölçeği ile değerlendirildi, 4 hastada hafif zeka geriliği varken bir tanesinde de donuk zeka vardı. Onbir hasta aynı zamanda Bender Gestalt görsel motor testi ile değerlendirildi ve 7 hastada görsel algılamada bozulma izlendi. Okuma hızı ve yazma testi okuma yazması olan 9 hastaya uygulandı ve 7 hastada okuma hızı daha yavaşken 6 tanesinde de yazma becerisi bozukluğu tespit edildi. Tartışma: Uzun zamandan beri benign çocukluk çağı parsiyel epilepsilerinde nörolojik ve nörofizyolojik bozukluğun olmaması ön koşul olarak düşünülmektedir. Ancak sadece yayınlanan bir kaç çalışmada iyi huylu çocukluk çağı oksipital epilepsilerinde hastaların bilişsel bulgularından bahsedilmiştir. Bu çalışmada iyi huylu çocukluk çağı oksipital epilepsi hastalarında değişik derecelerde bilişsel disfonksiyon ve görsel algılamada bozulma gösterilmiştir. İyi huylu çocukluk çağı oksipital epilepsi hastası çocuklarda uygun müdahalelerin sağlanması için bilişsel fonksiyonlar değerlendirilmelidir.

Anahtar Kelimeler

Oksipital; Epilepsi; Nörokognisyon

Abstract

Aim: The aim of this study was to evaluate the cognitive and visual perceptive functions in children with childhood epilepsy with occipital paroxysms (CEOP). Material and Method: Hospital charts of children ages 1 to 18 years who admitted to pediatric neurology out-patient clinic between 2009 and 2010 were reviewed. Twenty one children with a diagnosis of CEOP were identified. Sixteen of these children who accepted to include the study were evaluated with neuropsychological tests. Results: Two of five patients who were evaluated with Denver developmental screening test were found to have lower scores than their reference standards. Remaining 11 patients were evaluated with Wechsler Scales of Intelligence tests, 4 were mildly mental retarded and 1 had null intelligence. Eleven patients were also evaluated with Bender Gestalt Visual Motor Test and 7of them had disturbances in visual perception. Reading speed and writing norm tests were applied to 9 literate patients and 7 of them showed slower reading ability and writing ability was found worse in 6 patients. Discussion: The absence of neurological and neuropsylogical deficits has long been considered as a prerequisite for diagnosis of benign childhood partial epilepsies. However, only a few studies describing the cognitive profile of patients with CEOP have been published. The present study has demonstrated that the patients with CEOP had varying degree of cognitive dysfunction and disturbance in visual perception. In order to provide appropriate intervention, cognitive functions should be assessed in children with CEOP.

Keywords

Occipital; Epilepsy; Neurocognition

DOI: 10.4328/JCAM.2344 Received: 13.07.2014 Accepted: 06.08.2014 Printed: 01.03.2016 J Clin Anal Med 2016;7(2): 184-7
Corresponding Author: Ebru Kolsal, Bakırköy Dr Sadi Konuk Eğitim ve Araştırma Hastanesi Çocuk Nöroloji Bölümü Bakırköy, İstanbul, Türkiye.
GSM: +905055366258 F.: +90 2125424491 E-Mail: ebkut@yahoo.com

Introduction

Benign focal epilepsy of childhood (BFEC) is the most common form of the epilepsy in children aged from 3 to 12 years [1]. Diagnostic criteria proposed for benign partial seizures include absence of neurologic or intellectual deficits, easy control with antiepileptic drugs, spontaneous remission in adolescence, and characteristic electroencephalography (EEG) patterns [1].

Childhood epilepsy with occipital paroxysms (CEOP) is a type of benign focal epilepsy of childhood which is classified by Gastaut in 1982 [2]. CEOP represents 2-7% of benign childhood focal seizures [3]. The ILAE recognized early-onset childhood epilepsy with occipital spikes (Panayiotopoulos type) and differentiated it from the later-onset occipital epilepsy (Gastaut type) [4].

Early-onset benign childhood occipital epilepsy presenting between 3 and 5 years of age is characterized by the nocturnal seizures with tonic deviation of the eyes and vomiting [3]. Electroencephalography (EEG) reveals occipital or posterior temporal spikes with a distinctive morphology superimposed on normal background activity which does not seem related to clinical course and severity. Panayiotopoulos' syndrome is about half as frequent as benign focal epilepsy with central-temporal spikes. The frequency of seizures is generally low and prognosis is excellent with remissions occuring within few years from onset [3].

The Gastaut type has a later onset, presenting between 3 and 16 years, and seizures are characterized by visual symptoms rather than gaze deviation [5]. It has similar EEG abnormalities to that of early-onset type involving occipital spike wave activated by eye closure. However, the prognosis is somewhat poorer in terms of chance for remission.

One-third of patients with CEOP appear to have a mixed syndrome, with features of both Panayiotopoulos and Gastaut syndromes [6]. Thus they may represent a single clinical spectrum. Therefore we included children with both early-onset and late-onset occipital epilepsy syndromes in the present study.

Benign epilepsy with centrotemporal spikes (BECTS) is the most common benign focal epilepsy of childhood [1]. Most studies evaluated neuropsychological profiles of children with BECTS, and demostrated that children with benign focal epilepsy of childhood (BFEC) had varied cognitive difficulties [7,8].

However, limited data are available for the neurocognitive profiles of children with CEOP. While Gülgönen et al showed lower performance IQ score in children with CEOP [9], Germona et al. demonstrated normal performance but lower verbal IQ [10]. CEOP may theoretically affect visual perceptive function. Germano et al found reduced reading, writing and calculation abilities which were associated with visual memory and graph motor skills[10]. In the present study we aimed to investigate the cognitive and visual perceptive functions in patients with CEOP.

Material and Method

Subjects

Hospital charts of children ages 1 to 18 years who admitted to pediatric neurology out-patient clinic at Gazi University Medical Faculty Hospital between 2009 and 2010 were reviewed. Children with a diagnosis of CEOP were identified. Children those with progressive systemic or neurologic disorders like neurodegenerative or neurometabolic diseases, those with symptomatic occipital epilepsy who had any lesions in neuroimaging studies and those with concurrent psychiatric disorder other than ADHD were excluded from the study. Twenty-one patients with a diagnosis of CEOP were identified, and 16 of them who had a complete neuropsychological evaluation were included into the study. All children were developmentally normal.

Detailed information including demographic characteristics, seizure type, epileptic syndrome, age at onset, selected AED, duration of AED therapy and neuroimaging results were collected. Seizure types and epilepsy syndromes (Panoyitopoulus or Gestaut type) were classified according to the recommendations of the International League Against Epilepsy (ILAE) [4]. The electroencephalography findings of all patients were interpreted by a child neurology resident and a senior pediatric epileptologist.

Neuropsychological Evaluation

DENVER II (1992) is a revision and update of the Denver Developmental Screening Test, DDST (1967) [11]. Both were designed for use by the clinician, teacher, or other early childhood professional to monitor the development of infants and preschool-aged children. Clinician can identify children whose development deviates significantly from that of other children warranting further investigation to determine if a problem exists. The tests cover four general functions: personal social (such as smiling), fine motor adaptive (such as grasping and drawing), language (such as combining words), and gross motor (such as walking). Ages covered by the tests range from birth to six years. This method was standardized for Turkish children in 1999 [12].

Wechsler Intelligence Scale for Children (WISC-R): It consists of a test of two sub-scales that can be combined to form a total intelligence quotient (T-IQ) score that measures general intelligence performance. The two sub-scales include the following: verbal, and intelligence performance sections. The verbal intelligence quotient (V-IQ) scale was divided into: information, similarities, arithmetic, vocabulary, comprehension, and digit span. The performance intelligence quotient (P-IQ) scale was divided into: Picture completion, picture arrangement, block design, object assembly, coding, and mazes. Each sub-scale was considered independent, and data from the sub-scales were compared with data from the reference tables in the manual for each V-IQ, P-IQ, and T-IQ score calculation. T-IQ scores were classified in the following seven levels: ≥130, very superior; 120-129, superior; 110-119, normal-brilliant; 90-109, normal; 80-89, normal-awkward; 70-79, border-line; and ≤69, mental retardation.

The method was first developed by Wechsler in 1949, revised in 1986 and adapted to Turkey by Savaşır and Şahin in 1986 [13]. The two subtests' viability constants are 0.97 for verbal section, 0.93 for performance section and 0.97 for the overall viability .

Bender Gestalt Test: It is a psychological assessment instrument used to evaluate visual-motor functioning and visual perception skills in both children and adults. It is the most frequently used test with task to draw figures [14-16].This test is also used to measure short term memory, working memory capacity, visual perception and visual configuration skills. Scores on the test are used to identify possible organic brain damage and the degree maturation of the nervous system ("organic-

ity") [17]. Main function of the occipital lobe is processing the visual perception. Occipital lobe also provides the perception of colours, movement and visual experience.

Reading speed and writing norms test: This test was developed by Erden and friends [18]. A total of 2572 children attending the 1st-5th grades of randomly selected elementary schools belonging to different socioeconomic levels constituted the sample of the study in Turkey. The number of words read from grade-level appropriate scripts in 1 minute indicated reading speed. Reading speed was found respectively 45, 73, 91, 97and 120 words per minute for 1st to 5th grade elemantary school children. A three-sentence script containing repeating consonants p-b-t-d-m-n-v-f was dictated to determine writing errors. Mean, standard deviation and standard error scores indicate that objective academic achievement criteria were reached [18].

Denver developmental screening Test was applied to patients under the age of 6 years and WISC-R, Bender Gestalt Visual Motor Test and reading writing test which is compatible with age was performed to older patients.

Results

A total of 16 children consisting of 6 (37.5%) females and 10 (62.5%) males were enrolled in the study. Mean age at clinical evaluation was 9 years with a range from 3 to17 years.

Overall, 12 (74.9%) patients had complex partial seizures and remaining 4 (25.1%) patients had simple partial seizures. One of the patients experienced visual symptoms as the sole ictal event of simple partial seizure. The EEGs revealed clear occipital epileptiform activity in all children, a left parietooccipital discharge during sleep in 3, a right parietooccipital discharge during sleep in 5, and bilateral parietooccipital discharges in 8 patients. Five patients had a diagnosis of concurrent hyperactivity and attention deficit disorder. All patient underwent brain MRI studies which were all normal. Patients anti epileptic drug therapy were 6 with valproic acid, 1 oxicarbazepin, 1 carbamazepin, 3 with phenobarbital and 1 with topiramate.

Five patients who were younger than the age of six were evaluated with Denver Developmental Screening Test and 2 of them were found to have lower scores than their age standards. Remaining 11 patients were evaluated with Wechsler Scales of Intelligence Tests (WISC-R) and 6 of them were found to have normal intelligence, 4 were found to have mild mental retardation and 1 was found to have null intelligence. Minimum and maximum scores of WISC-R Test are shown in table 1. These 11 patients were also evaluated with Bender Gestalt Visual Motor Test and 7 (63.6%) of them were found to have disturbances in visual perception.

Table 1. WISC-R Intelligence scores

	Minimum	Maximum	Mean
Verbal Intelligence Score	51	100	79,6923
Performance Intelligence Score	54	104	81,7692
Total Intelligence Score	58	101	80,0000

Reading speed and writing norm test was applied to 9 literate patients and 7 children showed slower reading ability as compared to reference scores (Table2). When compared to reference scores, writing ability was found worse in 6 patients, of them, 5 mixed the letters and 1 skipped the letters. (Table 3)

Table 2. Patients' reading ability

	Patient(n)	Percentage(%)
Compatible with age	2	22,2
Incompatible with age	7	77,8
Total	9	100

Table 3. Patients' writing ability

	Patient (n)	Percentage (%)
Normal	3	33,3
Mixing Letter	5	55,6
Skipping Letter	1	11,1
Total	9	100,0

Consequently nine children had normal intellectual efficiency (IE) or psychomotor development whereas the other exhibited slight mental retardation (6) or severe delay (1). Cognitive deficits were nevertheless found in children with normal IE. Bender Gestalt Visual Motor Test exhibited visual perception disturbances in the majority of patient (7/9). Moreover, reading speed ability was lower in 7/9 writing tests pointed out errors in 6/9.

Discussion

The absence of neurological and neuropsylogical deficits has long been considered as a prerequisite for diagnosis of benign childhood partial epilepsies. However several studies have demonstrated subtle cognitive dysfunction in children with benign focal epilepsy of childhood, particularly in patients with benign childhood epilepsy with centrotemporal spikes [7,8]. Deonna et al studied the cognitive profiles of the patients with BFEC and found mild, varied and transient cognitive difficulties [8]. Gündüz et al also observed minor motor deficit and language problems in patients with BFEC [7]. Some other studies showed impairment in attention, visual motor organization, short term memory, auditory verbal memory and learning abilities at patients with BFEC [19, 20].

Moreover, with psychometric testing, almost half of the children with epilepsy met criteria for a learning disability. However, only a few studies describing the cognitive profile of patients with CEOP have been published [9, 10, 21]. The present study has demonstrated that half of the patients with CEOP had varying degree of cognitive dysfunction. Similar to our results, Gülgönen et al found lower performance IQ in CEOP patients [9]. Although statistical insignificant, Polat et al also found lower performance IQ in patient with occipital epilepsy [21].

Benign childhood epilepsy with occipital paroxysms has been found to be associated with selective dysfunction in perceptive-visual attentional ability, verbal and visual-spatial memory abilities, visual perception, visual-motor integration, some language tasks, reading, writing abilities, and arithmetic abilities [10]. Similarly, 7 of 11 patients evaluated with Bender Gestalt Visual Motor Test showed disturbances in visual perception in our series, and reading and writing abilities were worse as compared to reference scores. A previous study demonstrated disturbances in Bender-Gestalt Test especially for symptomatic epilepsy group [21]. Another study also showed disturbance in

visual perception but no difference between symptomatic and non symptomatic groups [23]. In our series, all patients had nonsymptomatic CEOP and more than half of them showed visual perceptive disturbance.

It is unclear whether the seizures or antiepileptic drugs cause learning disability, or the underlying neurologic pathology causes both seizures and abnormalities in perception, memory, and visual-motor skills. Also we couldn't find any relation between drugs and neurophyschologic tests. Children with benign childhood epilepsy with centrotemporal spikes have been demonstrated as having difficulties in memory and phonologic processing skills [23]. The risk of reading disability and speech sound disorder were found increased in siblings of rolandic epilepsy patients, suggesting that they are independently inherited traits rather than a consequence of the epilepsy itself [24] .

It is well known that children with epilepsy demonstrate an excess incidence of inattention and impulsivity. In a recent study, 70% of children with benign epilepsy were found to have attention-deficit hyperactivity disorder [25]. Similarly, 5 of 16 patients in our series had a diagnosis of concurrent hyperactivity and attention deficit disorder.

Limitations

One of the limitations of our study was that we were not able to form a control group. Instead we used age appropriate reference scores for comparisons. The small sample size of the study subjects was the other limitation of the study. Antiepileptic drugs may influence the cognitive function in children with epilepsy. The exact pathogenic mechanisms underlying the association between epilepsy itself, antiepileptic drugs and disturbed cognitive functions have not been fully elucidated. Because of ethical issue, we could not compare the results between patients received AEDs and patients untreated. Further comparative studies investigating the cognitive functions in children with CEOP who have not received AED therapy are needed.

Conclusion:

Children with CEOP may have cognitive dysfunction. In order to provide appropriate intervention, cognitive functions should be screened in children with CEOP.

Competing interests

The authors declare that they have no competing interests.

References

1. Holmes GL. Benign focal epilepsies of childhood. Epilepsia 1993;34 (Suppl 3):49–61.
2. Gastaut H. A new type of epilepsy: benign partial epilepsy of childhood with occipital spike-waves. Clin. Electroencephalogr 1982;13(1):13–22.
3. Panayiotopoulos CP, editor. A clinical guide to epileptic syndromes and their treatment. London: Springer; 2010.p.347-57.
4. Berg AT, Berkovic SF, Brodie MJ, et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. Epilepsia 2010;51(4):676–85.
5. Caraballo RH, Cersósimo RO, Fejerman N. Childhood occipital epilepsy of Gastaut: a study of 33 patients. Epilepsia 2008;49(2):288–97.
6. Taylor I, Berkovic SF, Kivity S, et al. Benign occipital epilepsies of childhood: clinical features and genetics. Brain 2008;131(9):2287–94.
7. Gündüz E, Demirbilek V, Korkmaz B. Benign rolandic epilepsy: neuropsychological findings. Seizure 1999;8(4):246–9.
8. Deonna T, Zesiger P, Davidoff V, Maeder M, Mayor C, Roulet E. Benign partial epilepsy of childhood: a longitudinal neuropsychological and EEG study of cognitive function. Dev Med Child Neurol 2000;42(9):595–603.
9. Gülgönen S, Demirbilek V, Korkmaz B, Dervent A, Townes BD. Neuropsychological functions in idiopathic occipital lobe epilepsy. Epilepsia 2000;41(4):405–11.
10. Germanò E1, Gagliano A, Magazù A, Sferro C, Calarese T, Mannarino E, et al. Benign childhood epilepsy with occipital paroxysms: neuropsychological findings. Epilepsy Res 2005;64(3):137–50.
11. Frankenburg WK, Dodds J, Archer P, Shapiro H, Bresnick B. The Denver II: a major revision and restandardization of the Denver Developmental Screening Test. Pediatrics 1992;89(1):91–7.
12. Anlar B, Yalaz K. Denver II Gelişimsel Tarama Testi. Türk çocuklarına uyarlanması ve standardizasyonu. Ankara: Hacettepe Üniversitesi Yayınları; 1996.
13. Savaşır I, Şahin N. Wechsler Çocuklar için Zekâ Ölçeği (WISC-R) El Kitabı. Ankara: Türk Psikologlar Derneği Yayınları; 1995.
14.Piotrowski C. A review of the clinical and research use of the Bender-Gestalt test. Percept Mot Skills 1995;81(2-3):1272-4.
15. Costenbader V, Allison MR, DiFonzo N. Kindergartenscreening: a survey of current practice. Psychol Sch 2000;37(4):323-32.
16. Cashel ML. Child and adolescent psychological assesment:current clinical practices and impact of managed care. Prof Psychol Res Pract 2002;33:446-53.
17. Bender L. A Visual Motor Gestalt Test and its clinical use (American Orthopsychiatric Association Research Monograph No. 3). New York Am. Orthopsychiatr. Assoc. 1938.
18. Erden G, Kurdoğlu F, Uslu R. İlköğretim okullarına devam eden Türk çocuklarının sınıf düzeylerine göre okuma hızı ve yazım hataları normlarının geliştirilmesi. Türk Psikiyatri Dergisi 2002;13(1):5-13.
19. Piccirilli M, D'Alessandro P, Sciarma T, Cantoni C, Dioguardi MS, Giuglietti M, et al. Attention problems in epilepsy: possible significance of the epileptogenic focus. Epilepsia 1994;35(5):1091–6.
20. Weglage J, Demsky A, Pietsch M, Kurlemann G. Neuropsychological, intellectual, and behavioral findings in patients with centrotemporal spikes with and without seizures. Dev Med Child Neurol 1997;39(10):646-51.
21. Polat M, Gokben S, Tosun A, Serdaroglu G, Tekgul H. Neurocognitive evaluation in children with occipital lobe epilepsy. Seizure 2012;21(4):241–4.
22. Piazzini A, Saetti C, Turner K, Fiorino A, Canger R, Canevini MP. Visuoperceptive impairment in adult patients with occipital lobe epilepsies. Epilepsy Behav 2009;15(2):256–9.
23. Northcott E1, Connolly AM, Berroya A, Sabaz M, McIntyre J, Christie J, et al. The neuropsychological and language profile of children with benign rolandic epilepsy. Epilepsia 2005;46(6):924–30.
24. Clarke T, Strug LJ, Murphy PL, Bali B, Carvalho J, Foster S, et al. High risk of reading disability and speech sound disorder in rolandic epilepsy families: case-control study. Epilepsia 2007;48(12):2258–65.
25. Bennett-Back O, Keren A, Zelnik N. Attention-deficit hyperactivity disorder in children with benign epilepsy and their siblings. Pediatr. Neurol 2011;44(3):187–92.

How to cite this article:
Kolsal E, Serdaroglu A, Soysal S, Gurkas E, Yılmaz U. Cognitive Findings in Benign Childhood Epilepsy with Occipital Paroxysms. J Clin Anal Med 2016;7(2): 184-7.